When are HPA-1b1b1b red cells needed?

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Vein to Vein 2015
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Objectives of this presentation

- Present a patient case where HPA-1b1b red cells were required.
- Provide a brief overview of Human Platelet Antigens.
- Provide an overview of Posttransfusion Purpura.
Patient M.L.

- 74 year old female
- 06/Jan/2015 – “lower extremity injury left hip”
- Type and Screen
  - A Rh positive
  - Antibody screen negative
  - Blood bank administrative data inquiry = HPA-1a negative. Can only receive cellular blood products from HPA-1a negative individuals.
- Hemoglobin = 149
- Platelet count = 212
Patient M.L.

- Family history of Posttransfusion Purpura (PTP)
- Two family members had PTP related deaths (intracranial hemorrhage)
- Platelet antigen testing performed on family members in 2000
- 29 family members typed
  - 9 = PL^{A2} (HPA-1a negative)
  - 20 = PL^{A1} (HPA-1a positive)
Posttransfusion Purpura (PTP)

- First described in 1959
- Characterized by thrombocytopenia 5 to 10 days after a blood transfusion in patients previously sensitized by pregnancy or transfusion
- Patient develops a potent platelet specific antibody usually to human platelet antigen (HPA)-1a
- PTP has also been associated with immunization to the following platelet antigens:
  - HPA–1b
  - HPA–3a
  - HPA–3b
  - HPA–4b
  - HPA–5b
  - HPA–5a
  - HPA–15b
33 HPAs expressed on six different platelet glycoproteins

Nomenclature adopted by the International Society of Blood Transfusion

Systems classified numerically according to date of publication and alphabetically to reflect their frequency in the population
Human Platelet Antigens

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<tr>
<th>Antigen</th>
<th>Traditional Name</th>
<th>Phenotypic Frequency</th>
<th>Glycoprotein (GP)</th>
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<td>GPIIIa</td>
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PTP - Incidence

- Incidence estimates between 1:50,000 and 1:100,000 transfusions
- Disease of adults
  - Youngest reported case 16 year old female
- Females outnumber males by a ratio of five to one
PTP – Clinical Presentation

- Purpuric skin lesions, mucous membrane hemorrhage, expistaxis, gastrointestinal bleeding, bleeding from urinary tract
- Mortality is caused by intracranial hemorrhage
- Severe thrombocytopenia occurs within one week following a blood transfusion
  - Red blood cells or whole blood usually precipitate PTP
  - Apheresis plasma, fresh frozen plasma or platelet concentrates can also trigger PTP
- Patients may experience transfusion reactions before and/or after PTP manifestation
PTP - Mechanism

- Antibody involved in PTP is an alloantibody that behaves like an autoantibody destroying autologous and allogeneic platelets
- 3 theories
  - Immune complexes bind to platelets through the Fc receptor causing destruction of platelets
  - Patient’s platelets adsorb a soluble platelet antigen from donor plasma making them susceptible to immune destruction
  - Platelet alloantibody has auto reactivity that develops when patient is re-exposed to a foreign platelet specific antigen
Laboratory tests show severe thrombocytopenia

Platelet count of less than 10

Bone marrow aspirates show normal or increased megakaryocytes

Platelet antibody assays
  - Usually detect an serum antibody with HPA–1a specificity

Typing patient’s platelets
PTP - Treatment

- Platelet transfusions not effective
- Duration of thrombocytopenia in untreated patients = 2 weeks
- Corticosteroids
  - High dose steroids (2mg prednisone per kg)
  - Normalization of platelet count within 1 week
- Plasma exchange with FFP
  - Responses occur after 12.5 days
- IVIG
  - First line therapy
  - 1g/kg/day for 2 days
  - Recovery occurs 3.7 days
PTP - Prognosis

- Most patients recover spontaneously 7 to 48 days after onset of thrombocytopenia
- Treated with IVIG, patients respond within 4 days of treatment
- PTP does not usually recur after subsequent transfusion
- Patients with documented history of PTP should receive
  - Antigen negative blood products
  - Autologous red cells
  - Platelet typing of family members to find suitable blood donors
  - Use of washed or frozen/deglycerolized is questionable
    - 2 reports of PTP from such products
Patient M.L.

- 10/Jan/2015 – surgery; fractured femur
- Hemoglobin = 121
- Platelet count = 174
- Crossmatch - 4 red cell units requested
  - 2 HPA-1b1b red cell units
    - Both red cell units collected 08/Jan/2015 from apheresis platelet donors
      - Group O Rh positive (Calgary)
      - Group A Rh negative (Edmonton)
  - 1 HPA-1b1b apheresis platelet
    - Group A Rh positive
Patient M.L.

- Post surgery 11/Jan/2015
  - Hemoglobin = 98
  - Platelet count = 169

- Patient discharged 15/Jan/2015
  - Hemoglobin = 96
  - Platelet count = 241

- No blood products required
  - Both HPA-1b1b red cell units returned to CBS 19/Jan/2015
The following comments were added to the blood bank administrative data inquiry file for this patient and her HPA-1b1b family members:

- Family history of PTP.
- Contact TM (transfusion medicine) physician for transfusion requirements.
References

- Popovsky MA. Transfusion Reactions. 4th ed.
- Curtis BR, McFarland JG. Human Platelet Antigens. Vox Sang. 2013
Questions